

## Idiopathic pulmonary vein thrombosis?

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### Keywords

CT scan, idiopathic, pulmonary vein thrombus.

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### Abstract

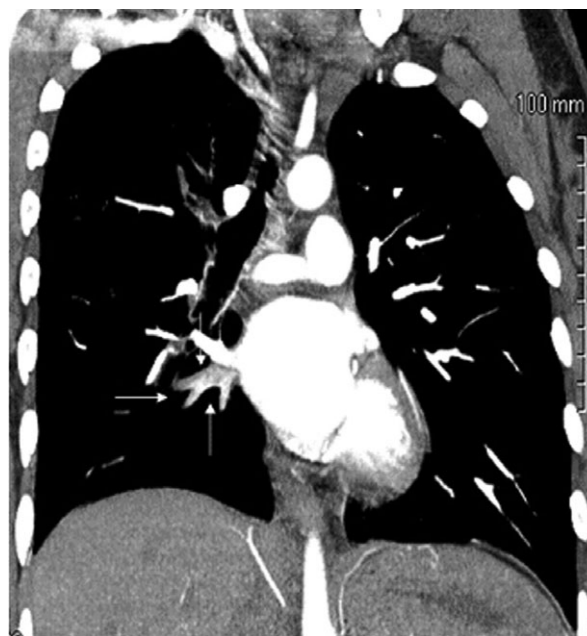
Idiopathic pulmonary vein thrombosis (PVT) is a rare disease which is likely under-diagnosed because of nebulous presentations. Accurate diagnosis is essential to prevent complications.

## Introduction

We describe a patient with pulmonary vein thrombosis (PVT) who presented with non-specific chest pain, evaluated with computed tomography (CT) angiography, and discuss the limited cases in the literature, providing guidance for clinicians.

## Case Report

A 26-year-old female gravida 5 para 5 presented to the emergency room with a single complaint of pleuritic chest pain. She denied shortness of breath or exertional dyspnoea. Past medical history was unremarkable. Chest pain began one week previously and progressed in severity over three days before presentation. Afebrile, she denied complaints of infection, rash, cough, haemoptysis, leg swelling, or contact with the sick. No illicit drug use, smoking, or regular alcohol consumption was reported. No family history of vasculitis or clotting disorders was noted. Physical examination revealed normal vitals with normal heart and lung sounds. Extremities exhibited no rashes, Homan's sign, or petechiae. The remaining examination was unremarkable. Haemogram revealed hypochromic microcytic anaemia and thyroid stimulating



**Figure 1.** The CT angiography (coronal plane) reveals a well-defined filling defect with partial occlusion (arrows) in the right inferior pulmonary vein.



**Figure 2.** The CT angiography revealing a well-defined filling defect with partial occlusion (arrows) within the right inferior pulmonary vein. Cardiac chambers are labelled.

hormone (TSH) of 10.24 (range 0.35–5.00 uIU/mL). Pulmonary function tests were normal.

Chest X-ray showed bilateral infiltrations in lower lobes in an interstitial pattern, inconsistent with pneumonic processes. Chest angiogram (Figs. 1–2) revealed right hilar mass or lymph node causing encasement of the right main pulmonary artery and infiltrates in the right middle and lower lobes, consistent with PVT. Positron emission tomography showed metabolically active focal consolidation in right middle lobe at lung periphery; standardized uptake (SU) value was 4.7. Serologic workup for vasculitis and connective tissue disorders and thrombophilia panel were all negative. ANA, lupus anticoagulant, and C-ANCA were all negative. Erythrocyte sedimentation rate (ESR) was slightly elevated, 49 (range 0–20 mm/h by manual technique), and P-ANCA was 25 AU/mL (range 0–19 AU/mL). Electro-navigational bronchoscopy with EBUS biopsy was negative for malignancy, lymphoma, endobronchial tumours, and granulomatous process. Small blood vessel biopsy was negative for vasculitis.

The patient was started on oral anticoagulation. After discharge, the patient had no difficulties. However, repeat CT scan after two months post discharge demonstrated residual narrowing of the right pulmonary vein. Oral anticoagulation with weekly monitoring of INR was continued. The patient was subsequently lost to follow-up.

## Discussion

Idiopathic PVT is a very challenging diagnostic entity because of non-specific presentations and limited image modalities. Known aetiologies of PVT include lung cancer; postoperative complications after lobectomy and lung transplantation; fibrosing mediastinitis; hypercoagulable states, such as protein C and S deficiencies; and Factor V Leiden mutations. [1] Only seven cases of idiopathic PVT

have been reported, including the current case (see Table 1). [2–6] Males and females appear equally affected; mean age in the series is 48, the current case representing the youngest in the series. Symptoms are non-specific and include dyspnoea, cough, haemoptysis, chest pain, and fever. Although touted as idiopathic, several patients had an underlying myocardial infarction and another had elevated homocysteine levels. Our patient exhibited positive P-ANCA. No predilection was noted for thrombus location. Delayed presentation or diagnosis was associated with lobectomy. Oral anti-coagulation appears to be the mainstay of treatment. A high level of suspicion is needed to confirm the diagnosis.

Imaging modalities of choice are pulmonary angiography (CTA), ventilation-perfusion scan, transesophageal echocardiography (TEE), and cardiac gated magnetic resonance imaging (MRI) which can also distinguish bland from tumour thrombus [2]. TEE is more accurate for distal pulmonary veins. Hurwitz et al. [7] have suggested treatment with antibiotics to prevent complications associated with thrombosis and have also showed that collaterals play a major role in the presentation of symptoms.

Thrombi that are small and non-occlusive can be treated with systemic anticoagulants. Antibiotics are also necessary due to the high risk of secondary infection of infarcted tissue [8,9]. Symptomatic patients or those with large thrombus should be treated with thrombolytics or surgical thrombectomy [8]. The current patient was empirically treated for pneumonia with antibiotics and warfarin was started after diagnosis. Duration of anticoagulation is always questioned. Some patients may initially present with complications, such as pulmonary gangrene, peripheral embolization, and myocardial ischaemia [4].

In conclusion, idiopathic PVT is a rare disease with ambiguous presentations. PVT is challenging to diagnose and treat. A detailed evaluation including risk factors, complications, and benefits of intervention appears to best serve treatment. Idiopathic PVT complications in literature reported cases include delayed diagnosis and gangrenous lung, transient ischemic attacks, stroke, peripheral embolization and chronic lung conditions. While definitive treatment of idiopathic PVT is unknown given its rarity, lung consolidation and pulmonary gangrene may require antibiotics. Anticoagulation may prevent clot propagation, embolization, TIA, and stroke. Accurate diagnosis is essential to prevent debilitating and fatal complications.

## Disclosure Statements

Appropriate written informed consent was obtained for publication of this case report and accompanying images.

**Table 1. Literature reported idiopathic pulmonary vein thrombosis.**

Author, year	Age/ sex	Symptoms/presentation	Past medical history	Imaging	Location	Hypercoagulability	Treatment(s)
Selvidge SD et al. [2]	33 F	Acute left sided abdominal pain, nausea and vomiting	Sickle cell anaemia	Contrast enhanced helical abdominal CT and confirmed with ECG-gated MRI	Right inferior pulmonary vein (RIPV) thrombosis extending to left atrium	Not reported	Oral anticoagulation but non-compliant. Two months later PVT still present but smaller
Alexander et al. [3]	47 F	Massive haemoptysis with three days of chest pain and dyspnoea. Complete consolidation of the LLL	Unremarkable	CT (high-resolution) scan. Intraoperative finding	Intraoperative finding. Left inferior pulmonary vein (LIPV)	Unknown	LLL lobectomy. No long-term treatment
Komatsu et al. [4]	57 M	Chest pain with myocardial infarction (MI)	Dyslipidaemia	CT chest, coronary angiogram by MDCT	Bilateral lower pulmonary veins	Negative work up	Antiplatelet for CAD and warfarin
Mumoli et al. [5]	80 M	Chest pain with MI and heart failure (EF-40%)	Dyslipidaemia	CTA	Left superior pulmonary vein	Homocysteinemia, otherwise negative	LMWH then to warfarin
Wu et al. [6]	30 M	Intermittent left chest pain for six months	Unremarkable	CT angiography (CTA) chest	Left inferior pulmonary vein thrombosis extending to atrium	Negative along with normal connective tissue tests	Left atrial mass resection and left lower lobectomy plus long-term anticoagulation
Rana et al., 2016	63 M	Sudden onset of central chest pain	Unremarkable	CTA chest, TEE	Pulmonary vein	Negative	Oral anticoagulation
Kollipara 2016 (current case)	26 F	Pleuritic chest pain	Gravida 5 para 5	CTA chest	Right inferior pulmonary vein	Positive for P-ANCA	Oral anticoagulation

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